

Women's Bleeding Disorders

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It's a simple fact: you can't treat what you don't acknowledge. When it comes to hemophilia, we don't acknowledge to what extent women are actually affected. Across this country, and the world, women are under-diagnosed and under-treated and, not surprisingly, that translates into women suffering and dying. For so many years, women who actually have hemophilia have instead been labeled as "symptomatic carriers." Proper diagnosis needs to be based on documented factor levels and not on gender when it comes to hemophilia, but setting aside two centuries of medical lore is tough. Nowadays, we know that "symptomatic carrier" is not appropriate or accurate. Newer studies have shown women labeled as symptomatic carriers have low factor levels; fifty percent of females who are "carriers" for hemophilia have Factor VIII or IX levels below 50%. Women may show permanent joint damage even with factor levels that would be considered adequate. The vast majority of women who have a son diagnosed with hemophilia, do not know their own factor levels. And if a woman believes she is "just" a carrier, she may end up accepting sub-optimal treatment by her health care providers, from routine procedures like dental work, to surgeries and trauma care.

How is it that so many health professionals, as well as the general public, still are under the misapprehension that hemophilia is visited only on males? Genetics is a complex arena of research. Everyone knows that males have an X and a Y chromosome as their 23rd pair and females have two X chromosomes. It's also commonly known that the gene for hemophilia A and B is carried on the X chromosome. What is lesser known, even by health professionals, is a situation called "Lyonization" or "X inactivation" in which women can actually have the "clotting" gene shut off, leaving the X chromosome with only the hemophilia gene "turned on." The percentage of cells in a woman's body affected by this Lyonization can vary, leaving some women with very low levels of clotting factor.

Women are subject to many types of bleeding disorders. Here are some examples:

VON WILLEBRAND DISEASE (VWD)

Officials estimate that 1-2 percent of the general population has von Willebrand disease (vWD) and is living undiagnosed, making vWD the most common bleeding disorder in the world. It is found equally in men and women, but again extremely under-diagnosed. Those with vWD may also have low factor VIII levels. Women can have deficiencies in other factors such as VII, XI (Hemophilia C), and XIII.

PLATELET DISORDERS

Platelet disorders are also a possibility when abnormal bleeding is present. Platelet disorders include Glanzmann's Thrombasthenia, Bernard-Soulier Syndrome, and Platelet Storage Pool Deficiency. Platelet disorders may involve low numbers, an inability to stick well to each other, or inability to travel to the site of the injury.

ABNORMAL UTERINE BLEEDING (AUB)

One common problem women, in and out of the bleeding disorder community, encounter is abnormal uterine bleeding. In the US, surveys report that 53 in 1000 women, ages 18-50 years of age, have had AUB. Abnormal uterine bleeding can be defined as bleeding for more than seven days, need to change pad or tampon every hour, passing clots larger than a quarter, blood loss greater than 80ml or daily activities limited due to heavy flow. AUB can impact women's quality of life, productivity, and utilization of healthcare services.

CONNECTIVE TISSUE DISORDERS

Women who have a connective tissue disorder such as Ehler's Danlos disease may have increased bleeding tendencies. They may have fragile blood vessels or loose joints that easily dislocate. This increases the chance of bleeding into soft tissues, muscles, and joints.

Do I Have a Bleeding Disorder?

In addition to AUB, a bleeding disorder shouldn't be ruled out if a woman experiences two or more of these symptoms:

- easy bruising (raised and larger than a grape in size)
- nosebleeds (frequent and or lasting longer than 10 minutes)
- prolonged bleeding after surgery or injury
- prolonged bleeding from dental work
- post-partum hemorrhage
- a family history of a bleeding disorder

When assessing a patient's bleeding disorder, it is important to ask about prior bleeding episodes and to characterize the type of bleeding.

- Bleeding into the skin and mucous membranes is associated with disorders of the platelets and blood vessels
- Bleeding into soft tissue, muscle and joints is associated with the presence of hemophilia or other disorders of coagulation proteins.

We encourage any woman who experiences unusual bleeding, or whose family history includes a bleeding disorder or suspected bleeding disorder, to get tested. The first step to treatment is understanding and acknowledging the condition. With your health, as with so many other aspects of your life, knowledge is power! ■■